

Brief Report

Anomalous origin of the circumflex coronary artery in a patient with atrial and ventricular septal defects

Abdullah Dogan, Omer Gedikli, Mehmet Ozaydin

Suleyman Demirel University, Sevket Demirel Heart Center, Isparta, Turkey

Abstract We report a patient in whom the circumflex coronary artery arose anomalously from the right aortic sinus. The patient also had atrial and ventricular septal defects in the setting of Eisenmenger physiology. We recommended transplantation of the heart and lungs.

Keywords: Coronary arterial anomalies; septal defects; anatomy; congenital cardiac disease

ACCORDING TO ANGIOGRAPHIC STUDIES, arterial anomalies are found in about 1% of all patients, with anomalous origin of the circumflex coronary artery being the most common anomaly.¹ The artery can arise abnormally from the right aortic sinus,^{2–5} and such anomalous origin may accompany other cardiac anomalies, including atrial septal defects or septal aneurysms.^{2,6} As far as we are aware, however, such anomalous origin of the circumflex artery has not been found previously in a patient with combined atrial and ventricular septal defects.

Case report

A 31-year-old-male presented with effort dyspnoea and angina. He had been known to have a ventricular septal defect since the age of 10 years. When seen during this presentation, his lips and tongue were cyanotic. A loud second heart sound, and a systolic murmur, was heard at the left sternal border. The electrocardiogram showed features consistent with right ventricular hypertrophy, while the chest X-ray revealed global cardiac enlargement, a prominent pulmonary infundibulum, and indirect evidence of pulmonary hypertension. His haematocrit was measured at 60%. Cross-sectional echocardiography showed large atrial and ventricular septal defects,

right ventricular hypertrophy, dilated right side cardiac chambers, and moderate tricuspid valvar regurgitation. During catheterisation, the catheter was passed from the right to the left atrium through the atrial septal defect, and atrial angiography was performed. Saturations of oxygen in the right and left atriums were 62% and 85%, respectively. The saturation in the right ventricle was 52%, and pressures of 130/0/12 mmHg were measured. The left ventricle was entered with catheter through the ventricular septal defect. The saturation of oxygen in this chamber was 80%, with pressures measured at 120/0/12 mmHg. Ventriculography confirmed the existence of the septal defect. Saturations and pressures in the pulmonary arteries were 53% and 130/80/93 mmHg, respectively. After giving intrapulmonary nitroglycerin at a dose of 300 µg, and verapamil at 250 µg, the pulmonary arterial pressures decreased to 120/65/84 mmHg. By the oxymetric method, we calculated the pulmonary-to-systemic shunt to be 0.7, and the ratio of pulmonary to systemic vascular resistances of 1.2. We then performed coronary angiography. This showed no luminal narrowing, but revealed that the left coronary artery, arising from the left coronary aortic sinus, divided into anterior interventricular and intermediate branches (Fig. 1), but that the circumflex artery arose from the right aortic sinus, passing behind the aorta to reach the left atrioventricular groove (Fig. 2). Both the circumflex and intermediate arteries provided nutrient branches to the superior and inferolateral aspects of the left ventricle. The patient was referred for heart–lung transplantation.

Correspondence to: Abdullah Dogan MD, Yayla Mh, Ismet Pasa Cd, 1533 Sk. No: 1/10, 32100 Isparta, Turkey. Tel: +90 246 2238616; Fax: +90 246 2327542; E-mail: adogan35@hotmail.com

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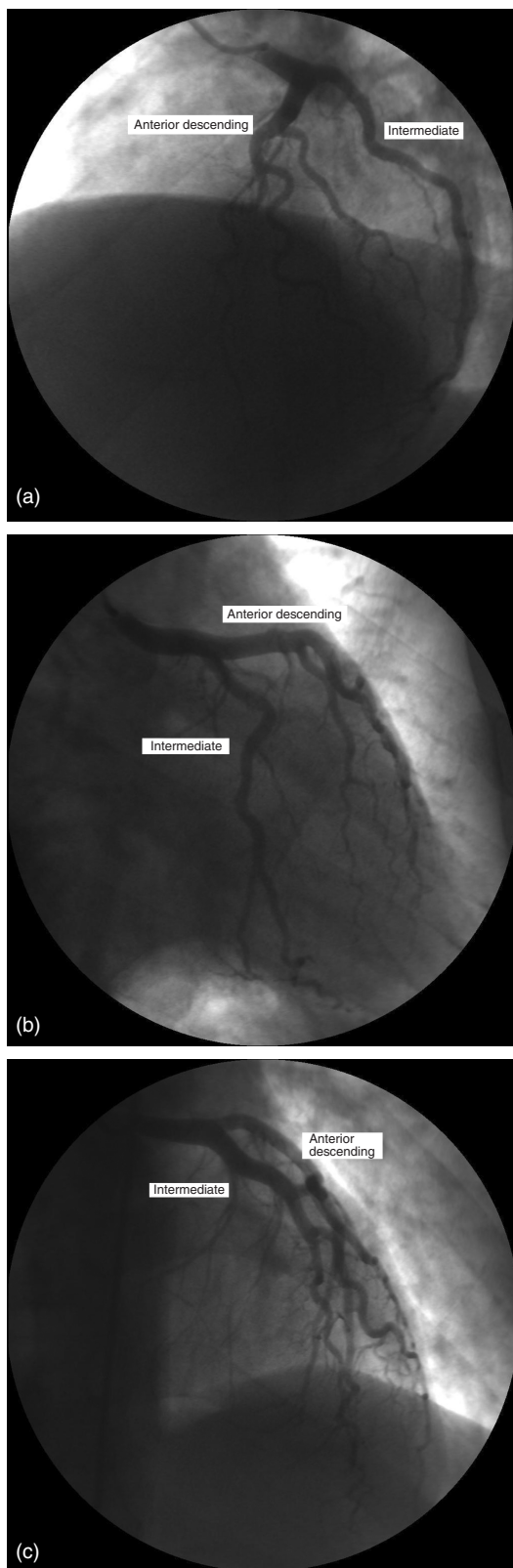


Figure 1.
 Division of the main stem of the left coronary artery into anterior descending and intermediate arteries, as shown in left (a) and right anterior oblique views (b), and in antero-posterior projection (c). No circumflex artery is seen passing into the left atrioventricular groove.

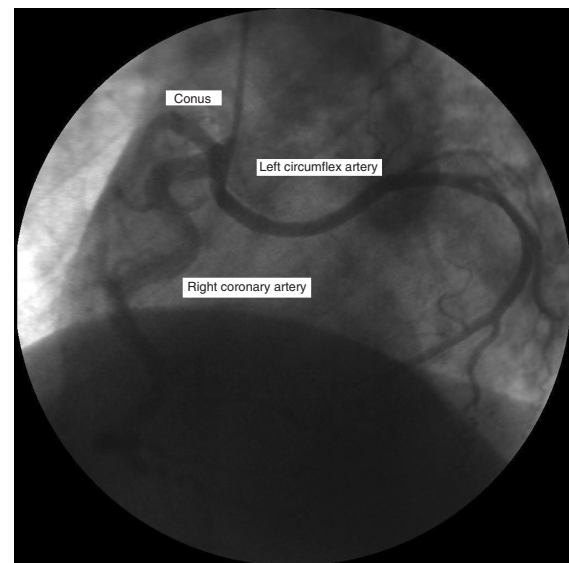


Figure 2.
 As shown in the left anterior oblique view, the right coronary artery, its infundibular branch, and an anomalous circumflex artery all take origin from the right aortic sinus. The circumflex artery passes through the transverse sinus behind the aorta to reach the left atrioventricular groove.

Discussion

The most common abnormal origin of the coronary arteries is for the anterior descending and circumflex arteries to arise separately from the left coronary aortic sinus, followed by origin of the circumflex artery from the right aortic sinus or right coronary artery.¹ In about one-third of normal patients, however, the main stem of the left coronary artery trifurcates rather than bifurcating, with an intermediate artery arising between the anterior descending and circumflex arteries.⁷ In our patient, the left coronary artery continued to give rise to the anterior interventricular and intermediate branches. Thus, there was dual supply to the oblique margin of the left ventricle from both the intermediate and anomalously arising circumflex arteries.

Anomalous origin of the circumflex artery from the right aortic sinus may be found incidentally, or can be seen with other cardiac anomalies, such as atrial septal defect or atrial septal aneurysm.^{2,6} Dual circumflex arteries have been reported twice. In the first description, the arteries arose from the left main stem and also from the aorta.⁸ In the second report, the arteries were said to arise from the left main stem and the right coronary artery,⁹ but the arterial arrangement in this case was comparable to our own, and hence we question whether the artery arising from the main stem should be described as a circumflex artery, since it passes directly to the obtuse margin of the left ventricle rather than coursing in the

atrioventricular groove. No associated anomalies were described in these reports.

Although anomalous origin of the coronary arteries is usually considered a benign finding, sometimes they can result in myocardial ischemia or infarction.^{1,3,5} Moreover, the circumflex artery, when originating from the right aortic sinus, is said to be more likely to be atherosclerotic.³ In our patient, however, coronary arteriography proved the absence of atherosclerosis. Our patient, however, had defects of both the atrial and ventricular septums. Such defects may accompany coronary arterial anomalies, including a single artery from the right aortic sinus,⁶ or origin of the right coronary artery from the pulmonary trunk.¹⁰ To our knowledge, however, the combination as seen in our patient has not previously been described.

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